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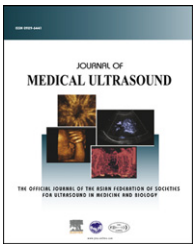

CASE REPORT

Dilated Cardiomyopathy: An Infrequent Cause of Posthepatic Portal Hypertension

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KEY WORDS
color Doppler ultrasound,
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Dilated cardiomyopathy is an infrequently encountered cause of posthepatic portal hypertension. In this case study, a patient presented with dilated cardiomyopathy with features of portal hypertension. Dilated portal and splenic veins with bidirectional flow, multiple portosystemic, and pelvic collaterals were observed. Grossly dilated inferior vena cava and hepatic veins with prominent right atrial pulsatality variations were also observed. While cardiomyopathy is described in literature as one of the causes of posthepatic portal hypertension, it is seldom documented.

Introduction

Portal hypertension is commonly caused by cirrhosis worldwide [1,2]. Dilated cardiomyopathy is an infrequent cause of portal hypertension and portosystemic collaterals [3]. Ultrasound with color and pulsed Doppler is the primary imaging modality of choice in evaluation of portal hypertension. Spectral wave analysis helps in evaluating the direction of flow and velocities in portal and hepatic veins [4,5]. In the case of a cardiac cause there will be pulsatile flow in the portal veins and altered waveforms in hepatic veins [6—8]. Dilated cardiomyopathy is one of the less common causes of portal hypertension and to our knowledge has not been documented as a cause of portosystemic collateral formation.

Case report

A 48-year-old female patient presented with complaints of dyspnea, abdominal distention, and pedal edema for 6 months, which had worsened in the past 2 weeks. On examination the patient had abdominal distention, bilateral pitting pedal edema, and a mildly enlarged liver. Laboratory investigations showed altered liver parameters (elevated transaminases and reduced hemoglobin). On grayscale ultrasound, the patient had a mildly enlarged

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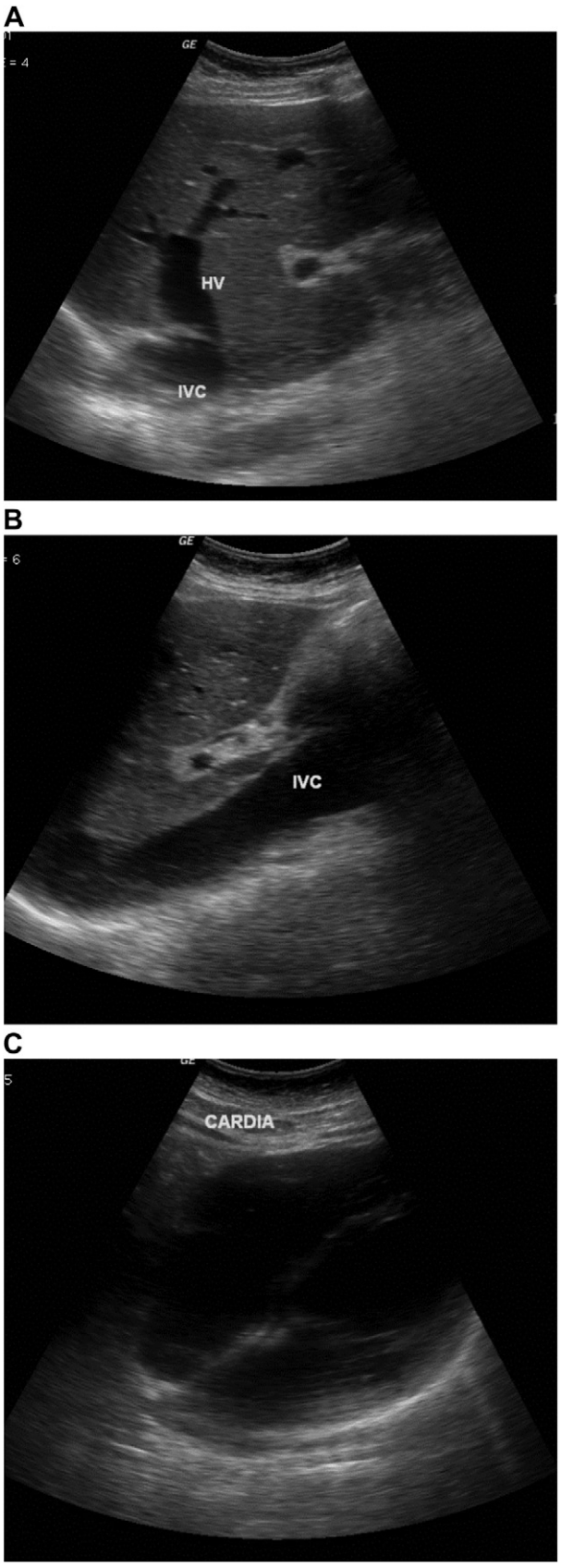

Fig. 1 (A) Dilated inferior vena cava (IVC) and middle hepatic vein (HV) at the junction of these veins. (B) Dilated intrahepatic portion of IVC. (C) Dilatation of all chambers of the heart with thinning of the myocardium.

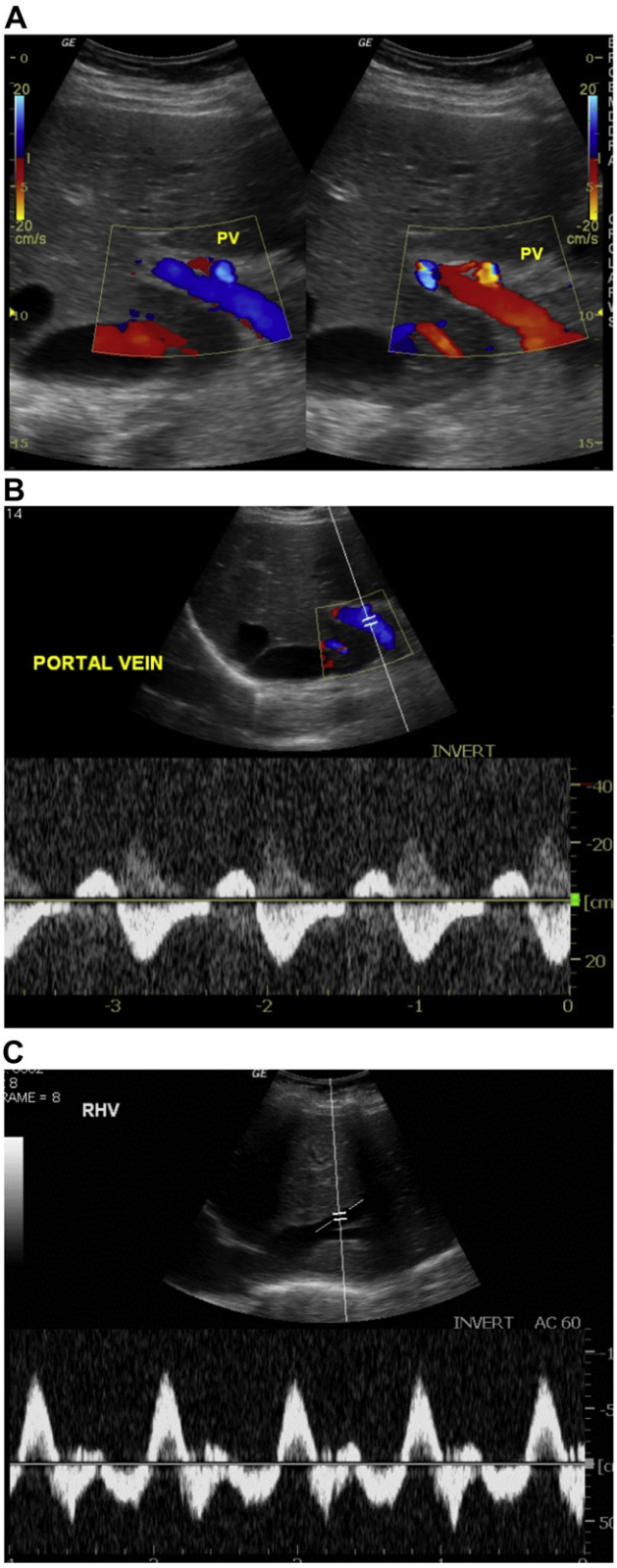

Fig. 2 (A) Bidirectional flow is seen in the portal vein (PV) on color Doppler. (B) Pulsatile flow is seen in the portal vein on pulse-wave Doppler imaging. (C) In tricuspid regurgitation, prominent *a* and *v* waves are observed in the hepatic veins. RHV = right hepatic vein.

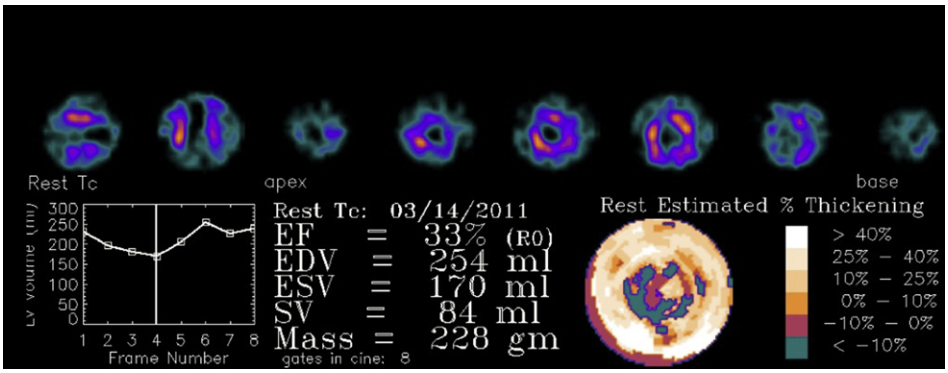

Fig. 3 Single-photon emission computed tomography showing reduced ejection fraction and ischemic changes. EF = ejection fraction; EDV = end diastolic volume; ESV = end systolic volume; SV = stroke volume.

liver measuring 17.5 cm which was normal in echo texture. The portal vein and spleen were normal in size. There were multiple dilated periportal and perisplenic collaterals, and no thrombus within the portal or splenic veins. The hepatic veins and inferior vena cava were dilated with no identifiable thrombus or obstruction (Fig. 1A and B). The patient had moderate ascites (Fig. 1C). There was dilatation of all chambers of the heart and reduction in ejection fraction, but no significant hypertrophy of the cardiac muscles or mural thrombus was seen. On color and pulse Doppler imaging, the portal vein showed forward and reverse flow with mild reduction in forward flow velocities (Fig. 2A and B). The superior mesenteric and splenic veins also showed the same type of spectral pattern consistent with right atrial pulsatility variation. The hepatic veins showed abnormally high *a* and *v* waves with small *s* and *d* waves (Fig. 2C). Cardiac single-photon emission computed tomography was then performed, which showed decreased left ventricular function with ejection fraction of 33% (Fig. 3). Also, infarct was seen in the left anterior descending artery territory and ischemic changes were seen in the inferior, anteroseptal, and lateral walls. In addition to the above findings there were dilated pelvic collaterals.

Discussion

Portal hypertension can be caused by a myriad of conditions [1,2]. They can be broadly classified into prehepatic, hepatic, and posthepatic causes. Hepatic causes can be further subclassified into presinusoidal, sinusoidal, and postsinusoidal [1,2]. Among the causes of portal hypertension, cirrhosis is the most common cause worldwide. Posthepatic conditions are not very common as compared to hepatic and prehepatic causes. The common causes of posthepatic portal hypertension are inferior vena cava obstruction, constrictive pericarditis, tricuspid regurgitation, Budd—Chiari syndrome, and arterial—portal venous fistula [3]. One of the infrequent causes of portal hypertension with multiple portosystemic collaterals is dilated cardiomyopathy. Cardiac causes are generally overlooked as a cause of portal hypertension. In a patient with normal echo texture of liver, and when the possibility of portal venous or splenic vein thrombosis has been eliminated, one should always rule out an underlying cardiac cause. Cardiomyopathy is a common cardiac diagnosis that may result as a consequence of a variety of pathologies, both acquired and congenital [9—11].

Although rare, dilated cardiomyopathy is one of the neglected causes for portal hypertension [6]. To our knowledge this is one of the first cases to be published where dilated cardiomyopathy has caused such extensive portosystemic collaterals. Grayscale ultrasound imaging is very useful in evaluating cases of portal hypertension. It is used to assess the causes of portal hypertension and to rule out cirrhosis, thrombosis, collaterals, and other complications. In addition, color Doppler and pulsed-wave Doppler can be used to assess the waveform of portal vein, hepatic veins, and inferior vena cava [4,5]. Normal individuals typically demonstrate minimal variation in portal vein velocity on spectral Doppler analysis during breath holding [12]. The mildly phasic pattern is most likely to be a reflection of changes in transmitted right atrial pressures, with significant dampening by resistance in venules, sinusoids, and small portal vein branches. Pulsatile portal venous flow is said to be present when the minimal portal vein flow velocity drops to, or below, the baseline [6—8]. Causes that may result in a pulsatile portal venous flow include tricuspid regurgitation, aortic-right atrial fistula, or a fistula between portal and hepatic veins. Other ancillary findings in such cases include dilated IVC (diameter >2.5 cm) and hepatic veins with abnormal spectral waveform [13]. In addition to the above findings there were also dilated pelvic collaterals. A relationship between the pelvic collaterals and portal hypertension should also be considered.

Conclusion

In a patient with portal hypertension and dilated portosystemic collaterals in the absence of hepatic or prehepatic causes, cardiac causes should always be kept in mind. Dilated cardiomyopathy is a very rare cause of portal hypertension with multiple portosystemic collaterals.

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